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ORIGINAL ARTICLE

Outcome of early and late onset Fontan operation in patients with univentricular heart repair

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KEYWORDS

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Abstract *Objective:* To evaluate our experience in the Fontan procedure comparing those below and above 6 years of age.
Methods: A review of our clinical database was conducted to identify the patients who received extracardiac Fontan between 2002 and 2010. All demographic, echocardiographic, surgical, haemodynamic and follow-up data were collected. The overall mortality was defined as death occurring from the time of surgery to the most recent follow-up. Early postoperative death was defined as death occurring during admission or within 30 days from the operation. Seventy-six patients with functionally univentricular hearts were included in the study. Patients were divided into two groups. Group A included patients who had received extracardiac Fontan at the age of 6 years or less, whereas group B included patients who had received extracardiac Fontan at an age of more than 6 years.
Results: The overall hospital mortality was 7.9% (10.2% in group A and 5.9% in group B). No statistically significant difference was seen between the two groups regarding the postoperative

Abbreviations: A SEPT, atrial septectomy; AV, atrioventricular; AVSD, atrioventricular septal defect; BT, Blalock taussing; BCPA, bilateral cavopulmonary anastomosis; BDG, bidirectional glenn; COP, cardiac output; DKS, Damus-Kaye-Stantel; DILV, double inlet left ventricle; DORV, double outlet left ventricle; ECMO, extra corporeal membrane oxygenation; INR, international normalized ratio; IVC, inferior vena cava; IVS, intact ventricular septum; LPA, left pulmonary artery; PA, pulmonary artery; PAB, pulmonary artery band; PLE, protein-losing enteropathy; PS, pulmonary stenosis; SVC, superior vena cava; TAPVD, total anomalous venous drainage; TCPC, total cavopulmonary connection

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mortality including thrombosis, stroke, chylothorax, bleeding, pericardial effusion, wound infection, serious postoperative arrhythmias and protein losing enteropathy. On the other hand the mechanical ventilation duration, duration of hospital and ICU stay, duration of the chest tubes and the postoperative saturation was not significant between the two groups.

Conclusions: The age of the patient at the time of Fontan surgery does not affect the results, in terms of both morbidity and mortality.

1. Introduction

Since the first report of a successful Fontan operation in 1971, this maneuver has been performed with various modifications in patients with single-ventricle physiology [1,2]. Among these modifications, total cavopulmonary connection (TCPC), introduced by de Leval and colleagues [3] has become a standard method for the Fontan procedure because it provides better venous hemodynamics [4] and is less arrhythmogenic [5] than the other Fontan modifications.

The management strategy for patients with a functional single ventricle has evolved into a sequence of staged procedures with a final goal of achieving normal volume and pressure work for the single ventricle and normal or near normal oxygen saturation of the systemic blood [6].

Although Fontan completion provides near-normal systemic oxygen saturation and reduces ventricular volume overload in patients with single-ventricular physiology, some preoperative factors affect the early and long-term outcomes of this procedure and age at the time of Fontan is among these factors [7]. Since the strategy now tends to introduce the Fontan procedure to younger patients, the influence of patients' age at the time of surgery on long-term results remains unclear [8]. The TCPC procedure carries a greater risk for the adult patient than for children because the adult functional ventricle usually presents with complications caused by long-term chronic hypoxia, ventricular volume overload, and increased venous pressure, such as arrhythmia, protein-losing enteropathy (PLE), pleural effusion, ventricular dysfunction, and limited exercise capacity [9]. In this study we evaluated our experience in the Fontan procedure comparing those below and above 6 years of age.

2. Patients and methods

A review of our clinical database was conducted to identify the patients who received extracardiac Fontan between 2002 and 2010. All demographic, echocardiographic, surgical, haemodynamic and follow-up data were collected. The overall mortality was defined as death occurring from the time of surgery to the most recent follow-up. Early postoperative death was defined as death occurring during admission or within 30 days from the operation. The Institutional Review Board approved the present retrospective study. Informed consent for retention and use of patient data for scientific purposes was routinely obtained at the same time as consent for the procedure.

Seventy-six patients with functionally univentricular hearts were included in the study. Demographic, anatomic and haemodynamic characteristics are shown in Tables 1 and 2.

Median age at extracardiac Fontan was 5.9 years (range 2–34 years). We used the median value of 6 years to divide the study population into two groups. Group A includes patients who had extracardiac Fontan at the age of 6 or less, whereas group B includes patients who had extracardiac Fontan at an age more than 6 years. After Fontan completion all patients were kept on warfarin therapy (plus aspirin) aiming to keep the international normalized ratio (INR) between 1.8 and 2.5.

2.1. Surgical techniques

Through a median sternotomy the heart, ascending aorta, pulmonary arteries, superior vena cava, and IVC were dissected out. After aorta and bicaval cannulation (with the IVC cannulated as close to the diaphragm as possible), cardiopulmonary bypass was instituted, the snare around the cannula in the IVC was tightened, and the SVC was clamped just proximal to its anastomosis with the right pulmonary artery. If there was no planned concomitant intracardiac procedure, the procedure was basically performed with the heart beating. If we had to make an atrial septectomy, we transected the inferior vena cava and carried out atrial septectomy under cross clamp. The heart was arrested with cold blood cardioplegic solution administered in the antegrade fashion, followed by the opening of the atrium and unroofing of the coronary sinus and atrial septectomy if indicated. The main PA stump was divided, and the cardiac stump was sutured closed if necessary. The IVC was divided from the atrium, the atrial stump was closed with a 4–0 Prolene suture (Ethicon, Inc, Somerville, NJ), and the IVC cuff was prepared for anastomosis with the conduit. Bovine jugular vein xenograft (Medtronic's Contegra, Medtronic, Inc, Minneapolis, Minn) was used for 47 patients; and Dacron tube vascular graft was used in 29 patients. The use of the contegra or Dacron tube was chosen by the cardiac surgeon and in most of the cases it was decided according to the availability in the hospital stock. The mean diameter of the conduits used was 17.74 ± 1.86 mm (range 12.0–22.0 mm) for the entire cohort. The fenestrations were made in 15 patients (19.7%). The mean bypass time was 140.61 ± 41.88 min. Cardioplegic arrest was used in 56 patients, with a mean duration of 87.3 ± 29.1 min. Preoperative diagnosis and procedures before Fontan are illustrated in Tables 2 and 3.

Statistical analysis was performed with SPSS statistical program (SPSS 19 Inc., Chicago, IL, USA). The Shapiro–Wilk normality test was used to assess normal distribution. Continuous variables with normal distribution were reported as the mean ± the standard error. Continuous data without normal distribution were reported as the median with ranges. Categorical data were presented as number and/or frequency.

3. Results

The overall hospital mortality was 6 (7.9%). Only one patient was in group B and the remaining 5 patients in group A. The

Table 1 Demographic, anatomic and hemodynamic characteristics.

Variable	Group A (n = 59)	Group B (n = 17)	P value
Age	4.02 ± 1.07	12.6 ± 7.7	< 0.0001
Sex (male)	39(66.1%)	5(29.4%)	0.008
Weight	14.5 ± 2.2	31.2 ± 15.1	< 0.0001
Single ventricle morphology:			
Right	19 (32.2%)	4(23.5%)	NS
Left	30(50.8%)	5(31.3%)	NS
Intermediated	10(16.9%)	7(41.2%)	0.049
Atrial pressure (mmHg)	7.2 ± 3.4	7.4 ± 4.1	NS
Mean pulmonary artery pressure (mmHg)	11.9 ± 3.7	11.3 ± 3.4	NS
Ventricular end-diastolic pressure (mmHg)	8.98 ± 2.6	8.94 ± 2.9	NS
Pulmonary vascular resistance (woods unit)	1.6 ± 0.76	1.8 ± 0.5	NS
Arterial oxygen saturation (%)	82.3 ± 6.8	78.9 ± 5.9	NS

Table 2 Preoperative diagnosis.

Variables	Group A (n = 59)	Group B (n = 17)	Total (n = 76)
Double inlet left ventricle	12(20.3%)	3(17.6%)	15(19.7%)
Tricuspid Atresia	10(16.9%)	4(23.5%)	14(18.4%)
Double outlet right ventricle	9(15.3%)	3(17.6%)	12(15.8%)
Unbalanced AVSD	9(15.3%)	1(5.9%)	10(13.2%)
Dextrocardia, situs inversus with univentricular heart morphology	7(11.9%)	3(17.6%)	10(13.2%)
PA,IVS	5(8.5%)	1(5.9%)	6(7.9%)
Mitral atresia	5(8.5%)	1(5.9%)	6(7.9%)
Other complex anomalies*	2(3.4%)	1(5.9%)	3(3.9%)

* One case had Ebstien's anomaly of the tricuspid valve with pulmonary atresia, second case had Atrial inversion, multiple muscular VSDs, large ASD, large PDA, small TV and RV, the third case had dextrocardia, L-TGA, PS and sub PS.

Table 3 Procedures before Fontan.

Variables	Group A (n = 59)	Group B (n = 17)	Total (n = 76)
Bidirectional Glenn	12(20.3%)	5(29.4%)	17(22.4%)
BTS	2(3.4%)	0	2(2.6%)
BTS, BDG	15(25.4%)	8(47.1%)	23(30.3%)
PAB	2(3.4%)	0	2(2.6%)
BDG, PAB, A SEPT	19(32.2%)	0	19(25.0%)
DKS, BDG	3(5.1%)	1(5.9%)	4(5.3%)
KAWASHIMA	3(5.1%)	1(5.9%)	4(5.3%)
PAB, A SEPT	1(1.7%)	0	1(1.3%)

first patient in group A was 4 years old, with a diagnosis of unbalanced atrioventricular canal defect, malposed great arteries, pulmonary stenosis, double outlet right ventricle, and bilateral SVC. This patient had BTS, atrial septectomy, and bidirectional Glenn, she developed low cardiac output just few hours after the operation and died 5 days after the Fontan. The second patient was 6 years old, with diagnosis of dextrocardia, double inlet left ventricle, situs ambigus, interrupted IVC, pulmonary stenosis, and hypoplastic right ventricle. The patient had Kawashima, Aortic tear during redo sternotomy which was managed with total circulatory arrest. The patient was transferred to the ICU with open chest, but he had post operative severe bleeding that needed exploration. Unfortunately the patient did not recover. The third patient was 3 years old, had double outlet right ventricle, mitral atresia. This patient underwent arch repair with pulmonary artery banding followed by DKS and bidirectional Glenn. Unfortunately this patient was lost after 4 days due to aspiration in the ICU. The fourth patient was 3.5 years with a double outlet right ventricle, infundibular subvalvular pulmonary stenosis, malposed great arteries, and bilateral SVC had TCPC then developed low cardiac output connected to the ECMO for 7 days, died on ECMO support due to severe bleeding and multiorgan failure. The fifth patient was 1.3 years who was diagnosed as having an unbalanced atrioventricular canal defect, Bilateral SVC, TAPVD, PS. The plan was bilateral bidirectional Glenn. However, intraoperatively we found that the left SVC was very short and cannot be connected to do left Glenn, so intraoperative decision was taken to proceed for right bidirectional Glenn plus the Fontan (single stage total cavopulmonary connection) since he has favorable cardiac catheterization numbers. The patient developed low COP and expired after 3 days in the ICU. Retrospectively, the case was discussed in the mortality meeting and the group felt that

the patient age was not favorable for Fontan surgery although the cardiac catheterization numbers were good. The policy of the department was changed after that to do Fontan surgery only after the age of 3 years. The patient of group B was 7 years old with a diagnosis of dextrocardia, atrioventricular canal defect, hypoplastic left ventricle, and left pulmonary stenosis who was a foreign patient that had bidirectional Glenn in another hospital (outside the country) and was referred to our institution for Fontan completion. Although the patient was fully evaluated by echocardiography and diagnostic cardiac catheterization preoperatively, TAPVC to IVC was missed and it was discovered intraoperatively. The surgeon had to repair the TAPVC and to do the Fontan in the same setting. The patient developed low COP and expired in the ICU. Late mortality was reported in three patients all of them from group A. The first was 4.9 years diagnosed as having an unbalanced atrioventricular canal defect, Bilateral SVC, pulmonary stenosis, double outlet right ventricle had bilateral bidirectional Glenn, died in ER with vomiting and loose motions 27 months after Fontan with dehydration. The second was 2.5 years with a diagnosis of double outlet right ventricle, mitral atresia, atrial septal defect with a small left ventricle and had balloon atrial septostomy then DKS with Modified BT shunt, then the Glenn shunt, developed protein losing enteropathy that was not controlled and the patient died 6 months after Fontan. The third was 2 years and was diagnosed as having mitral atresia, double outlet right ventricle had balloon atrial septostomy, followed with the Glenn shunt with pulmonary artery banding and atrial septectomy. Later glenn was taken down and the patient had DKS with modified BT shunt. The patient then developed protein losing enteropathy. The patient had fenestration for the PLE but died in the ICU.

Five of our patients (all in group A, Table 3) had total cavopulmonary anastomosis as a single stage, one of them discussed in the mortality cases. The other four cases were charity patients who missed their Glenn stage in their countries and presented to our institute late. All of them had favorable cardiac catheterization numbers and all of them had a smooth postoperative course without any complications.

Complications following the Fontan procedure were thrombosis in 6 patients (10.2%) in group A and 2 patients (11.8%) in group B with no significance. In group A the first patient was 3 years with a dacron tube of size 18 who had mural thrombus immediately post operative that was managed with alteplase infusion in the ICU. The second was 5 years with contegra size 18 who had bilateral SVC with bilateral bidirectional Glenn then developed a small clot at the Fontan circuit that was managed with heparin infusion. The third patient was 4.5 Years with contegra size 18 who had clotting of the entire Fontan and proximal LPA. This patient was managed with alteplase infusion and was proved to have kinked contegra that was managed with 2 covered CP™ stents (NuMED Inc.) each 3.9 cm (inflated using size 18 balloon) in size. The fourth was 5 years with contegra size 18 who developed thrombosis 6 months after Fontan. This patient was managed with thrombectomy then connected to the ECMO circuit for 5 days. Later he was weaned from ECMO and discharged in good condition. The fifth was 3 years with contegra size 18 who developed thrombosis 12 months after Fontan and received alteplase. The last patient in this group was 5 years with contegra size 14 who developed thrombus at the wall of contegra that was managed with covered CP™ stent 4.5 cm (NuMED Inc.) that was inflated using a size 14 balloon. In group B the first patient was 9 years with a dacron tube of size 18 and developed thrombosed hepatic incorporation. He was managed with alteplase complicated by hemothorax. He needed to redo Fontan. The other patient was 7 years with contegra size 18 with thrombosis at proximal IVC and RPA that was managed with alteplase and heparin infusion. This patient also, needed stenting for kinked Fontan using single covered CP™ stent 4.5 cm (NuMED Inc.) that was inflated using a size 18 balloon.

Other complications were more in group A in the form of post operative pericardial effusion that was managed by drainage in a 5 year old patient with a dacron tube of size 16. Sternal wound infection that needed readmission was encountered in only one patient 2 years old with Contegra size 18. Two patients had serious post operative arrhythmias, the first one was 3.5 years with Contegra size 18 who developed AV block, and needed permanent pacemaker insertion. And the other one is 34 years with Contegra size 22 who developed SVT. Protein loosing enteropathy (PLE) was reported in only two patients one of them 4.5 years with contegra size18 who was diagnosed as Dextrocardia, DILV, DORV. He had modified BT shunt, then bidirectional Glenn. After the Fontan circuit he developed generalized edema. His total protein and albumin level were low. Diagnostic cardiac catheterization showed LPA stenosis with peak systolic pressure gradient 45 mmHg. Also his Fontan circuit was hypertensive with a mean pressure gradient of 28 mmHg. The patient had an LPA stent after which the peak pressure gradient dropped in the LPA to 23 mmHg and the mean Fontan pressure dropped to 17 mmHg. In addition, the patient received intensive diuretic therapy and subcutaneous low molecular weight heparin. The PLE resolved after several weeks of treatment. The other patient was mentioned with the mortality (Table 4).

4. Discussion

After Fontan and Baudet [1] reported a new operation for tricuspid atresia in 1971, various modifications of the Fontan procedure have been used for patients with a functional univentricular heart. The original atrio-pulmonary connection has evolved to the lateral tunnel total cavo-pulmonary connection (TCPC) to improve fluid dynamics and reduce energy losses, as demonstrated by de Leval et al. [3], and toward the concept of an extracardiac inferior vena cava to pulmonary artery connection, as proposed by Marcelletti et al. in 1990 [10].

Although it is known that pulmonary vascular development and maturation are essential conditions for a successful cavopulmonary connection, the timing of the surgical staging has been set arbitrarily, particularly the transition from superior cavopulmonary connection to Fontan. If this transition can be accomplished at an early age, the effects of persistent cyanosis and the potential for paradoxical embolization can be minimized [8].

There is a general consensus among clinicians regarding the age at which BCPA should be performed [11]. However, unlike BCPA, no consensus exists about the optimal timing of the Fontan operation. Some centers advocate early intervention to minimize the effects of persistent cyanosis and the risk of paradoxical embolism [8]. However, very young infants have shown higher values of pulmonary vascular resistance, lower oxygen saturation and a higher prevalence of arrhythmias after TCPC than older ones [12]. Furthermore, several groups demonstrated that TCPC cannot be considered a definitive

Table 4 Morbidity and mortality.

Variable	Group A (n = 59)	Group B (n = 17)	P value
Thrombosis	6(10.2%)	2(11.8%)	0.606
Stroke	2(3.4%)	2(11.8%)	0.219
Chylothorax	16(27.1%)	5(29.4%)	0.538
Bleeding	4(6.8%	0(0%)	0.355
Pericardial effusion	1(1.7%)	0(0%)	0.625
Sternal wound infection	1(1.7%)	0(0%)	0.625
Serious post operative arrhythmias	1(1.7%)	1(5.9%)	0.249
Protein loosing enteropathy	2(3.4%)	0(0%)	0.355
Mortality	6(10.2%)	1(5.9%)	0.506

Table 5 Post operative variables.

Variable	Group A (n = 59)	Group B (n = 17)	P value
Mechanical Ventilation (days)	1.88 ± 2.38	1.71 ± 1.69	0.780
Chest Tube Drainage (days)	10.81 ± 6.64	13.00 ± 9.23	0.280
ICU stay	2.67 ± 2.95	3.06 ± 2.14	0.618
Hospital stay	17.86 ± 13.38	21.24 ± 13.36	0.364
Post operative saturation	94.53 ± 3.43	93.47 ± 3.86	0.283

solution because of progressive, long-term attrition in terms of mortality and morbidity [13–14].

The data reported here demonstrate that Fontan completion can be successfully accomplished before the age of 6 years regardless of ventricular morphology. (Table 1). However, for performing Fontan in patients with low body weight, the size of the conduit, which lacks growth potential, might pose difficulties in the future [15].

Our results go with the results obtained by Pizarro et al. in 2006 [8], they reported that single-ventricle patients can transition toward Fontan completion as early as 1 year of age without additional morbidity or mortality. Fontan at a later age, and its outcome are similar. Additionally, their data support the concept that a favorable outcome after Fontan can be achieved regardless of the anatomic substrate, which goes with our data as well (Tables 2 and 3).

However, Kirklin and coworkers [16] reported 102 patients with a variety of cardiac malformations who underwent the Fontan operation between 1975 and 1985. Age younger than 4 years was a risk factor for early postoperative mortality in the early experience, but not with a more recent date of operation. A study from the Boston Children's Hospital reviewed 500 patients undergoing Fontan completion between 1973 and 1991, and also found that age <4 years at the time of the Fontan operation was a risk factor for failure. Although age at the time of Fontan has gradually decreased to about 2–4 years in some centers [10], when it comes to the timing of the Fontan, age is still considered, and this transition continues to rest on arbitrary timelines. Another reason frequently used to justify early intervention is that early TCPC completion avoids the effects of prolonged cyanosis. However, delaying TCPC completion does not necessarily translate into accepting long-standing suboptimal arterial saturation levels [10].

Indeed, as institutional policy, all patients in whom arterial oxygen saturation dropped below 80% and had no treatable cause of cyanosis (such as veno—venous collaterals) were immediately considered for TCPC completion. Moreover, all patients reported normal social life and attended school without gross neuro-developmental limitations at follow-up. However, some patients were operated on before the age of 6 although their saturations were higher than 80%, that is because these patients had adequate body weight (more than 15 kg) and we felt that the Fontan surgery should be done to avoid volume overload of their single ventricle.

In contrast to the implementation of early repair among patients amenable to biventricular physiology, the timing commonly used to consider Fontan completion delays the restoration of arterial oxygen saturation to near normal levels, and exposes these patients to the effects of persistent cyanosis and the potential for paradoxical embolization [17,18].

We found that there was no statistically significant difference between the two groups regarding the postoperative mortality including thrombosis, stroke, chylothorax, bleeding, pericardial effusion, wound infection, serious postoperative arrhythmias and protein losing enteropathy. Although 6 patients (out of 59) expired in group A, while only one patient (out of 17) expired in group B, the mortality rate between the two groups was not statistically significant (Table 4). Similarly, the mechanical ventilation duration, duration of hospital and ICU stay, duration of the chest tubes and the postoperative saturation was not significant between the two groups (Table 5).

Our data are supported by the results obtained by Ikai et al. [15] in their study, no difference in the duration of pleural drainage was noted between the groups. However the hospital stay in our patients was 17.86 ± 13.38 in group A and 21.24 ± 13.36 in group B (Table 5) which was less than the hospital stay reported in their study. They recognized that the hospital stay was considerably longer than that mentioned in any other previously published reports [15].

5. Conclusion

From the revision of our experience, the age of the patient at the time of the Fontan surgery does not affect the results, in terms of both morbidity and mortality.

Conflict of interest

None.

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